

# Transposition of the great arteries associated with a double left ventricular outflow tract<sup>1</sup>

R. H. KINSLEY, S. E. LEVIN, AND T. G. O'DONOVAN

*From the Departments of Cardio-Thoracic Surgery and Paediatrics, University of the Witwatersrand, Johannesburg, South Africa*

**SUMMARY** A case is described in which, at semilunar valve level, the aorta and pulmonary artery arose from inappropriate ventricles. Despite this, the outflow tracts to both vessels originated from the left ventricle. Embryologically, it is speculated that this anomaly is the result of normal rotation of the proximal conus, without concomitant truncal inversion, and excessive leftward shift of the proximal conus and conal septum or anterior and rightward deviation of the anterior segment of the ventricular septum. Surgical repair using a double conduit between the right ventricle and pulmonary artery and left ventricle and aorta, respectively, was unsuccessful.

Definition of specific outflow tract anomalies is often controversial. For example, transposition of the great arteries has variously been defined as a reversal of the anteroposterior relation of the semilunar valves (Goor and Edwards, 1973), any alteration in the position of the great arteries (Abbot, 1927), and the presence of mitral-aortic valve discontinuity as a result of a subaortic conus (Van Praagh *et al.*, 1967). Recently, several authors have favoured a more literal definition, redefining transposition of the great arteries to mean that both great arteries are 'placed across' the septum (trans-ponere means 'placed across') reversing the connection of the great arteries to the *morphological ventricle* (Van Praagh, 1973; Shinebourne *et al.*, 1976). Thus both great arteries arise from morphologically inappropriate ventricles. By the same token, double outlet ventricle is defined as a specific ventriculo-arterial connection in which more than one and a half great arteries arise from the same ventricular or outlet chamber (Kirklin *et al.*, 1973). We have accepted this literal definition of outflow tract anomalies.

However, according to the interpretation of embryological events by Goor and Lillehei (1975), the semilunar valves and outflow tracts of each of the major vessels are derived from different embryological segments. It is reasonable to anticipate, therefore, that the ventricle of origin of a major vessel (at semilunar valve level) may be at variance

with that of its outflow tract. In this communication a case will be described in which the left ventricle has a double outflow tract (aortic and pulmonary), but the origin of the great arteries at semilunar valve level is clearly from inappropriate morphological ventricles.

## Terminology

We have adopted the terminology recently proposed by Anderson *et al.* (1977) with regard to outflow tract anomalies. The term 'conus' (and its derivatives) is confined to embryological events. In the postnatal, or fully developed heart, the term 'infundibulum' is substituted for 'conus'. Thus the structure interposed between the two semilunar valves is described as the infundibular septum (parietal band of Van Praagh) (Van Praagh *et al.*, 1975). The muscle separating semilunar from atrio-ventricular valves is termed the ventriculo-infundibular fold regardless of whether it exists in the right or left ventricle. The trabecula septomarginalis (septal band of Van Praagh) (Van Praagh *et al.*, 1975) is an extensive right ventricular trabeculation with, superiorly, anterior and posterior limbs, reinforcing the anterior and inferior borders of many septal defects.

## Case report

A 5½-year-old boy had been cyanosed since birth. Chest x-ray film showed cardiomegaly and pulmonary plethora while the electrocardiogram

<sup>1</sup>Supported by a grant from the Stella and Paul Loewenstein Trust Cardiac Fund, University of the Witwatersrand.

showed a mean frontal QRS axis of  $+60^\circ$ , with evidence of right atrial and right ventricular enlargement. Small R waves were noted in the right chest lead. Cardiac catheterisation at age 6 days established the diagnosis of transposition of the great arteries, mild pulmonary stenosis, and a small ventricular septal defect. A Rashkind septostomy was performed with considerable improvement in oxygen saturation.

During the next 3 years increasing polycythaemia developed requiring multiple venesections. Cerebral thrombosis and hemiplegia occurred at age  $1\frac{1}{2}$  years with fairly good recovery. A Blalock-Taussig shunt was performed when the child was 4 years old, with subsequent improvement in oxygen saturation and fall in haematocrit. Recatheterisation at  $5\frac{1}{2}$  years confirmed the previous diagnoses. A systolic gradient of 41 mmHg was measured across the pulmonary outflow tract and the systemic oxygen saturation was 75 per cent. When the left ventricular systolic pressure was 70 mmHg, that in the right ventricle was 80 mmHg. Elective surgery was advised.

Using standard cardiopulmonary bypass techniques, a right ventriculotomy was performed and a 2 to 3 mm ventricular septal defect closed with a single horizontal mattress suture. A valved external conduit (16 mm) was then inserted between the right ventricle and right pulmonary artery. A similar conduit (18 mm) was placed between the apex of the left ventricle and left lateral wall of the ascending aorta. Initially, after discontinuing bypass, the haemodynamics appeared satisfactory. However, just before closing the chest, ventricular fibrillation occurred and resuscitation was not successful.

Necropsy confirmed ventricular non-inversion: the morphological right ventricle was right-sided and the morphological left ventricle was left-sided. The aortic valve was anterior, superior, and to the right, above the right ventricle. The pulmonary valve was to the left of, and slightly posterior to, the aortic valve. Though the pulmonary valve was overriding both ventricles (similar to the aorta in the normal heart), there was no direct communication between the right ventricle and the pulmonary valve or artery. Examination of the interior of the left ventricle disclosed two outflow tracts, separated by a muscular ridge (infundibular septum) approximately in a coronal plane (Fig. 1). The aortic outflow tract was dextro-posterior to the pulmonary outflow tract. The interior of the right ventricle (Fig. 2 and 3a) showed a ventricular septal defect 3 mm in diameter (the surgical suture was removed), triangular in shape, and immediately subaortic. It was bounded by the posterior limb of the trabecula septomarginalis, aortic valve, and, on the left, the

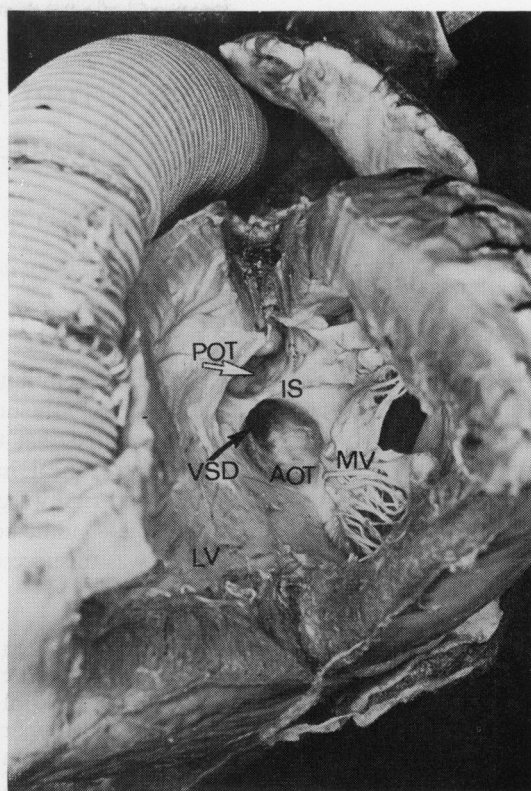


Fig. 1 Photograph of left ventricle (LV) with double (aortic and pulmonary) outflow tract. The left ventricle has been opened parallel to the left anterior descending coronary artery and immediately to the left of the left ventricular ascending aorta conduit. AOT, aortic outflow tract; IS, infundibular septum; MV, mitral valve; POT, pulmonary outflow tract; VSD, ventricular septal defect.

anterior ventricular septum. The posteroinferior rim of the defect was separated from the tricuspid valve by a 3 to 4 mm band of muscle. A well-developed ventriculo-infundibular fold separated the semilunar and mitral valves though it was only 2 to 3 mm wide below the aortic valve. The pulmonary outflow tract was mildly stenotic at its origin.

## Discussion

According to the observations of Goor and Lillehei (1975) and their interpretation of previous embryological descriptions, conotruncal inversion occurs in two stages. Firstly, the conoventricular junction (ostium bulbi) rotates  $110^\circ$  counterclockwise (looking downstream) during d-looping of the cardiac tube. This process transfers the proximal aortic

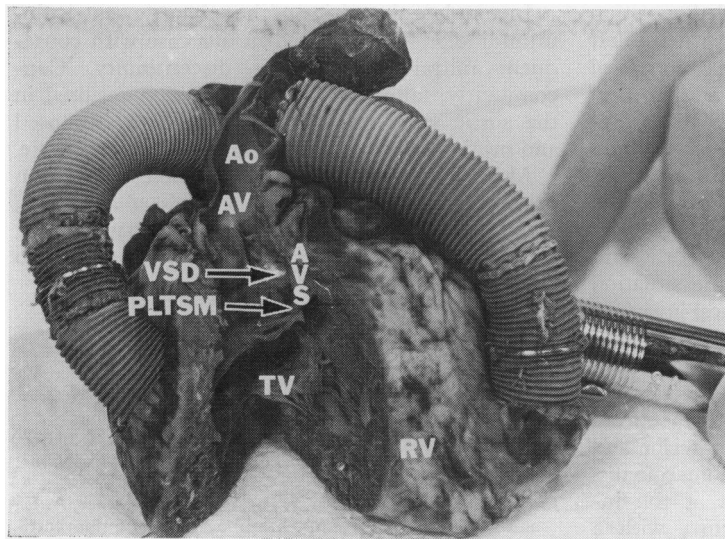


Fig. 2 Photograph of right ventricle (RV). The anterior wall of the right ventricle has been removed. The ventricular septal defect (VSD) is bounded by the posterior limb of the trabecula septomarginalis (PLTSM), aortic valve (AV), and anterior ventricular septum (AVS).

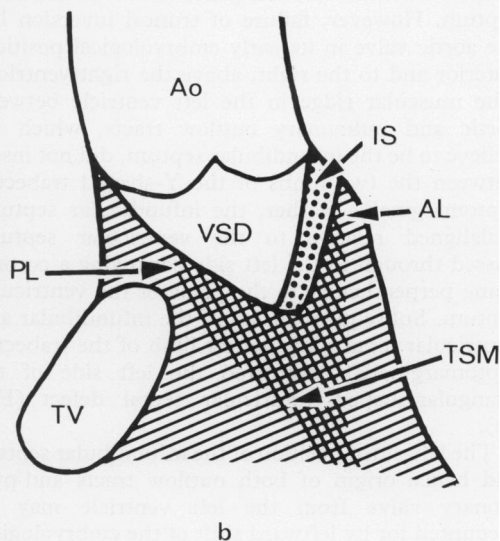
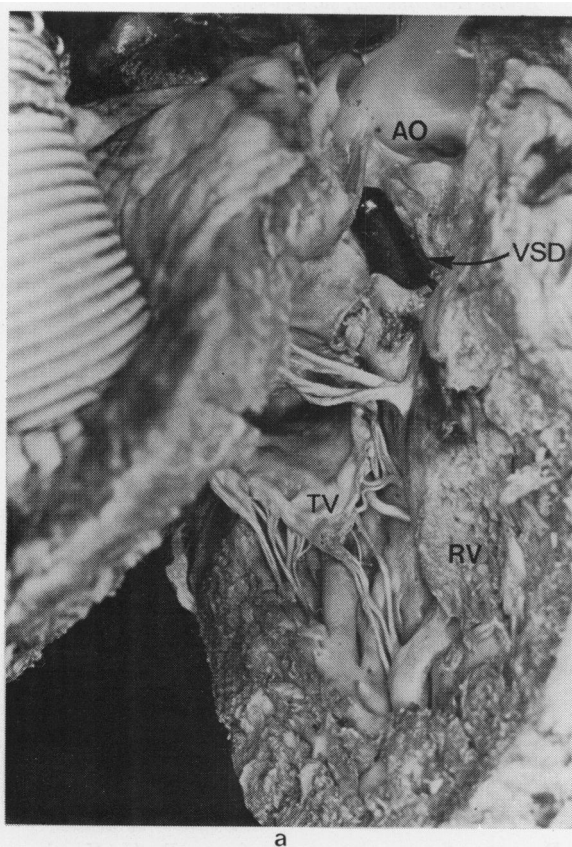


Fig. 3a Closer view of Fig. 2. Marker through VSD. AO, aorta; TV, tricuspid valve.

Fig. 3b Diagrammatic representation of relation between infundibular septum (IS) and trabecula septomarginalis (TSM). Ao, aorta; PL, posterior limb; AL, anterior limb; TV, tricuspid valve.

conus to a posterior (but still right-sided) position and enables it to establish contiguity with the left ventricular vestibulum. At this stage, the conotruncal ridges (destined to form the conal and truncal septa) occupy a spiral course. Secondly, but somewhat later, the truncus (including the developing semilunar valves) rotates in an identical manner. As a result, the conotruncal ridges are uncoiled, the aortic valve is transferred to the same posterior position as the aortic conus, and the aorta and pulmonary artery become entwined—the situation seen in the normal heart. Failure of truncal inversion results in dextroposition of the aortic valve (anterior and rightward displacement) and its origin predominantly, or entirely, from the right ventricle.

In the normal heart, the infundibular septum fills the Y-shaped space between the anterior and posterior limbs of the trabecula septomarginalis. Should this not occur, malalignment of the infundibular and ventricular septa occurs, with a resultant ventricular septal defect and a variety of outflow tract anomalies.

How does transposition occur with a double left ventricular outflow tract? Applying the embryological interpretation of Goor and Lillehei (1975), rotation of the ostium bulbi occurred in our case to account for the posterior position of the aortic outflow tract and coronal plane of the infundibular septum. However, failure of truncal inversion left the aortic valve in its early embryological position, anterior and to the right, above the right ventricle. The muscular ridge in the left ventricle between aortic and pulmonary outflow tracts, which we believe to be the infundibular septum, did not insert between the two limbs of the Y-shaped trabecula septomarginalis. Rather, the infundibular septum, malaligned relative to the ventricular septum, passed through to the left side occupying a coronal plane perpendicular to the plane of the ventricular septum. Subsequent fusion of the infundibular and ventricular septa (and anterior limb of the trabecula septomarginalis) completed the left side of the triangular-shaped ventricular septal defect (Fig. 3b).

The left-sided position of the infundibular septum and hence origin of both outflow tracts and pulmonary valve from the left ventricle may be accounted for by leftward shift of the embryological conotruncal junction (Goor and Edwards, 1973; Goor and Lillehei, 1975) or anterior and rightward deviation of the anterior segment of the ventricular septum (Anderson *et al.*, 1974). Though both processes were probably operative, we favour predominance of the latter, because of the relatively normal position of the pulmonary outflow tract and valve relative to the atrioventricular valves.

Incomplete absorption of the subaortic ventriculo-infundibular fold occurred in our case with consequent mitral-aortic fibrous discontinuity. Concomitantly, failure of truncal inversion resulted in the aortic valve and aorta remaining dextroposed and originating completely from the right ventricle.

Although 'posterior' transposition has been previously documented (Van Praagh *et al.*, 1971; Wilkinson *et al.*, 1975), our case is distinctive in that, at semilunar valve level, the aorta was anterior to the pulmonary artery.

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Requests for reprints to Professor R. H. Kinsley, Department of Cardio-Thoracic Surgery, University of the Witwatersrand, Hospital Hill, Johannesburg 2001, South Africa.